A SCALP NODULE IN AN INFANT

Fanfan S    Grove O    Laib A    Michie C.A.

ABSTRACT

Solitary, congenital scalp nodules, have a wide range of causes. This case report is followed by a literature review that explores a differential diagnosis of scalp nodules in a child. In this case the nodule represented an embryological anomaly, a neural tube defect. The relationship of such conditions to nutritional factors may be significant

KEYWORDS

Epidermoid Cyst, Dermoid Cyst, Encephalocele, Meningocele, Neural Tube Defect, Folic Acid

CASE REPORT

INTRODUCTION

A four-month-old boy was referred to a dermatology clinic in West London for a congenital mass on the vertex of his scalp. The nodule was firm, but not bony, had no discharge, no signs of infection or change in size since birth. No punctum was noted. The child had been born following an uncomplicated pregnancy and delivery. It was not clear if his mother had taken folic acid early in pregnancy or prior to his conception. Also noted, but assumed unrelated to the nodule, were 3 small café au lait spots (<0.5cm) and low-set ears. The spine and sacrum were normal, as was a careful neurological review including fundoscopy. Initial differential diagnosis included epidermoid cyst, pilar cyst, dermoid cyst or an encephalocele. This was the mother’s first child; there was no family history of congenital anomalies known.

An ultrasound review described a lobulated but fairly well-defined cyst within the posterior aspect of the scalp measuring 1.4cm x 0.9cm. The cyst had a tract through the cranium probably communicating with the dural space. No evidence of neural tissue was present within the cyst. An MRI showed, “a small fluid-filled soft tissue lesion in continuity with the intracranial contents through a bony defect with a fluid tract running adjacent to the falcine sulcus. This is most likely to represent a small meningocele or dermoid cyst with an intracranial connecting tract. No abnormality of the brain is evident”. The scalp lesion was excised and confirmed to be an atretic meningocele on
histological examination. The patient has recovered and shown normal growth, development and neurology since the procedure.

DISCUSSION

Scalp nodules occur frequently in children. The differential diagnosis is extensive including a variety of aetiologies such as congenital malformations (encephaloceles, nasal gliomas, dermoid cysts), acquired malignancies (sarcoma, Langerhans cell histiocytosis, metastatic neuroblastoma), infections, trauma or conditions with unclear causation such as epidermoid cysts (7). Neurofibromatosis in particular is associated with solid subcutaneous nodules and café au lait macules. Congenital, solitary, cystic nodules consistent with our case include only epidermoid cyst, pilar cyst, dermoid cyst, and encephalocele.

Epidermoid cysts and pilar cysts exhibit similar clinical presentation and histology. Histologically, both types of cysts consist of a wall of stratified squamous epithelium which produces keratohyalin granules, differentiated only by the pattern of keratinization (5). An epidermoid cyst most commonly presents as a compressible, non-fluctuant, solitary nodule with a shiny surface due to the upward pressure of the cyst. It is freely movable over the underlying tissues and size may range from less than 0.6 cm up to 5 cm (5,6). The presence of a punctum, which occurs due to blockage of the follicular aperture (often in association with acne vulgaris), may assist in identification
(5,6). Epidermoid cysts are most commonly found on the face, neck, and trunk, but may arise anywhere on the body. However, lesions of the scalp are more likely to be pilar cysts. In fact, 90% of pilar cysts develop on the scalp. Pilar cysts do not have an overlying punctum and are likely to be firmer and more mobile than epidermoid cysts. A cyst may be hybrid, showing characteristics of both epidermoid and pilar cysts (2). Epidermoid cysts are one of the most common benign skin nodules of adulthood but rarely present before puberty, so the earlier onset in this case is more suggestive of pilar cyst (5). Treatment for both types is surgical excision (2).

Congenital lesions made up of developmentally mature tissues, dermoid cysts, are usually non-pulsatile, cystic, firm and do not trans-illuminate. The cysts are often lined with keratinized squamous epithelium histologically (2). These hamartomatous lesions are thought to be a result of the sequestration of ectodermal tissues along the lines of embryonic segmental fusions during development. Depending on the location of the cyst it may contain hair, skin, fluid and glands (10). These benign lesions can be surrounded by hair although the skin covering it will usually demonstrate alopecia. Dermoid cysts frequently regress over time to form a scar (9). They range in size, usually several centimeters in diameter at the time of birth (10). Dermoid cysts of the scalp may have potential to develop an intracranial or intradural sinus tract resulting in a risk of infection of the CNS (10, 2) and therefore require surgical management.

In normal fetal development, the rostral neuropore is expected to close by the fourth week. Failure of complete closure of the cranium, known as neural tube defects, can result in protrusion of the meninges, brain tissue, or both from the skull. The resulting extracranial mass is referred to as a meningocele, unless it contains brain tissue in which case it is called an encephalocele (6). Such masses, or nodules, are often not detectable on routine antenatal ultrasound scans and may not be diagnosed until birth by their gross appearance (7). Common features of an encephalocele/meningocele are a dome shape, alopecia, “collar” of long hair surrounding the nodule, midline location, and occipital region (6,7,11). Neural tube defects have a multifactorial aetiology, including genetic causes and dietary deficiencies of folic acid or inositol (1). The exact mechanisms by which a layer of neural tissue persists intact in the cranial vault are not known. A number of studies have revealed higher rates of neural tube defects in West London, probably because of less effective implementation of antenatal guidelines in this poor urban area (3,4,12). Fortification of flour with folic acid has improved this specific problem in other countries, but is still under review in the UK (8).

CONCLUSION

A myelomenigocele can present as a scalp mass or nodule in infancy. The differential diagnosis of such a lesion is broad, but neural tube defects need to be excluded with appropriate imaging prior to surgical removal.
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REFERENCES